THE NERVOUS SYSTEM AND SPECIAL SENSES

The focus of this week’s lab will be pathology of the nervous system and special senses. A few representative cases have been chosen to illustrate some of the pathology of the nervous system and special senses.

There are two major types of stroke: ischemic and hemorrhagic. Ischemic stroke refers to loss of blood flow to the brain by a variety of mechanisms, most notably, thromboembolism. Meningitis can be caused by a number of organisms. In immunocompetent patients usually meningitis is of bacterial or viral origin, these can be differentiated by CSF findings and PCR for the responsible organism. In immunocompromised patients fungal meningitis can also occur.

The cases we will cover are:

A. Ischemic Stroke
B. Bacterial Meningitis
C. Amyotrophic Lateral Sclerosis
D. Retinoblastoma
E. Vestibular Schwannoma (Acoustic Neuroma)
A. ISCHEMIC STROKE

CC/HPI: A 35 year old woman known to have rheumatic mitral stenosis awakens in the morning to find the right side of her body paralyzed. She also complains of palpitations. She denies fever, neck stiffness, vomiting, headache, or transient ischemic attacks (TIAs).

PE: On physical exam the patient is not febrile, pulse is irregularly irregular. Right-sided hemiplegia; brisk reflexes on right side; right-sided Babinski present. Loud S1; apical mid-diastolic murmur and opening snap.

Labs/Imaging: EKG: indicates atrial fibrillation. Blood culture sterile; routine labs unremarkable; clotting time, bleeding time and PT normal. Echo: left atrial thrombus. CT, brain: infarct in posterior limb of left internal capsule.

Pathology: Normal brain (stained with H and E) is shown below:

Pathology: Autopsy findings from a more severe case (stained with H and E) is shown below:
Questions for everyone to consider:

Does this tissue look normal?

What is different about the sample from the patient?

Questions if you have been assigned this case:

What in this patient’s history increases her risk of stroke?

What medications are used to decrease risk of stroke in patients with atrial fibrillation?

What is tPA? How is it used to treat stroke? What are possible contraindications?

B. BACTERIAL MENINGITIS

CC/HPI: A 20 year old college student present with fever, headache, and neck stiffness that has been progressively worsening for the past two days. She has experienced some nausea and vomiting. Her friend that drove her to the hospital reports that she has been drowsy and difficult to arouse and noticed that she has developed a rash.

PE: On physical exam the patient is febrile. She is obtunded and confused. She has a non-blanching purpuric skin rash over her trunk and lower extremities. Nuchal and spinal rigidity. Kernig’s sign and Brudinzki’s sign positive. No focal neurological defects.

Labs: LP: elevated pressure; CSF is turbid, with 5,000 WBCs/mm³, glucose 35 mg/dl and total protein 500 mg/dl. Gram stain shown below. CT, brain: meningeal thickening and enhancement. Blood cultures
grew oxidase and catalase positive gram negative organism on chocolate agar; glucose and maltose metabolism also noted.

She was treated prophylactically with ceftriaxone pending blood cultures.

Pathology: A gram stain of this patient’s CSF is shown below:

Pathology: A autopsy finding from a similar patient reveals the following image:

Questions for everyone to consider:

Does this gram stain look normal?

If not, what is abnormal about it? What organism is likely to be the cause of her meningitis?
Questions if you have been assigned this case:

What is abnormal about the biopsy sample? What type of infection (bacterial, viral, fungal) is she likely to have based on this?

At what level is a lumbar puncture performed? Why?

What are the major causes of bacterial meningitis in the following populations: 1) neonates, 2) infants and young children, 3) adolescents and young adults, 4) older adults.

C. AMYOTROPHIC LATERAL SCLEROSIS

CC/HPI: A 49 year old man presents complaining of marked progressive weakness of his hand and arms, difficulty speaking, muscle wasting in his hands, and involuntary muscle contractions (fasciculations). He complains of dropping objects and has experienced difficulty with fine motor tasks (such as buttoning his shirts). He has no history of sensory symptoms, bladder or bowel dysfunction, fever, exanthema, dog bites, vaccinations, or spinal or cranial trauma.

PE: Lower motor neuron signs: bilateral wasting of hands, deep tendon reflexes absent in upper limbs, muscle weakness, fasciculations; upper motor neuron signs: positive Babinski sign, stiffness and spasticity of upper limbs; normal fundus, sensory system, and cranial nerves.


Pathology: The spinal cord form this patient is shown below (note: upper picture anterior view, bottom is posterior view):
Questions for everyone to consider:

Does this spinal cord look normal?

If not, what looks different about it (grossly and histologically)?

Questions if you have been assigned this case:

Is the area affected involved in relaying sensory or motor information?

How is this consistent with the patient’s symptoms? What findings suggest upper motor neuron damage and which suggest lower motor neuron damage?

What gene is associated with familial variants of this disease?
D. RETINOBLASTOMA

CC/HPI: A 2 year old child presents with her parents. Her parents report that she has had a decline in her vision particularly in her left eye and report that they think it is painful and tender. There is no familial history of similar disease in other family members.

PE: Fundoscopic exam reveals leukocoria (white pupillary reflex, cat’s eye reflex) in left eye (see image below). Strabismus also noted. Red reflex normal in right eye.

Labs/Imaging: All routine lab work normal.

Pathology: A normal eye (stained with H and E) is shown below:

Pathology: An eye from this patient (stained with H and E) is shown below; image on the right demonstrates Flexner-Wintersteiner rosettes:
Questions for everyone to consider:

Does this eye look normal?

If not, what structures appear altered?

Questions if you have been assigned this case:

What are the layers of the retina described in the atlas?

What gene mutation is associated with this condition?

What does the protein produced from this gene do?

E. VESTIBULAR SCHWANNOMA (ACOUSTIC NEUROMA)

CC/HPI: A 64 year old man complains of right sided hearing loss and intermittent unilateral tinnitus (ringing of the ear) and ear pressure. He also reports difficulty with balance and vertigo. His wife reports that he has been walking differently.

PE: Weber lateralizes to left ear. Rinne AC>BC bilaterally.

Labs/Imaging: Labs unremarkable. Contrast brain CT reveals 2.5 cm mass at cerebellopontine angle. Image is seen below.

Pathology: A brain CT from this patient is shown below:

Pathology: A brain from a patient with the same condition is shown below:
Pathology: After resection a sample from the tumor reveals the following images:
Questions for everyone to consider:

Does this brain look normal? If not where is the tumor?

What nerve is most likely affected in this patient? What two nerves does this nerve become that are seen in the atlas? Please be able to identify them.

Questions if you have been assigned this case:

What type of cells are seen in the histological images?

Explain the Weber and Rinne test findings.

What other nerves may be affected based on the position of this tumor?